King's College Hospital NHS Foundation Trust
KING'S HEALTH PARTNERS Pioneering better health for all
Guy's and St Thomas' NHS Foundation Trust

red cell *news*

A tri-annual newsletter for patients with sickle cell disease and thalassaemia

produced by the South Thames Sickle Cell & Thalassaemia Network

Issue 1 Summer 2012

WELCOME TO THE 1ST EDITION OF RED CELL NEWS

Hello and welcome to the 1st edition of red cell *news*. We plan to release three newsletters per year, Summer, Autumn/Winter and Spring.

red cell *news* is produced by the South Thames Sickle Cell and Thalassaemia Network (STSTN), a new collaboration of healthcare professionals involved in the care of people with sickle cell disease and thalassaemia.

This newsletter is produced with you, the patient and their families, in mind. So, if you want to get involved, get in touch with Annabelle (pictured right).

Send stories, photos, comments and any ideas about what you would like to see in this newsletter. Likewise, if you are a skilled graphic designer, photographer or illustrator, do get in touch.

You can volunteer your time by emailing Annabelle Kelly at: info@ststn.co.uk

Until the Autumn!

Annabelle Kelly
STSTN Support Manager

ALSO IN THIS ISSUE

ABOUT STSTN

The South Thames Sickle Cell and Thalassaemia Network (STSTN) is a new haemoglobinopathy collaboration led by Consultant Haematologists and Paediatricians at King's College Hospital, Evelina Children's Hospital and Guy's and St. Thomas' Hospital NHS Foundation Trusts.

The network also includes two district general hospitals, University Hospital Lewisham and the Queen Elizabeth Hospital Woolwich, with over 400 patients. In addition, Croydon University Hospital, St. George's Healthcare NHS Trust (a specialist tertiary centre for South West London and Surrey) and Darent Valley Hospital, feed into the network and have seen growing populations of patients with haemoglobin disorders.

The network also has strong links centres across South London and beyond, including Epsom & St. Helier Hospital, Medway Hospital and Brighton and Sussex University Hospital.

STSTN provides educational events and peer-support for consultants, medical trainees and other healthcare professionals involved in the care of patients with red blood cell disorders.

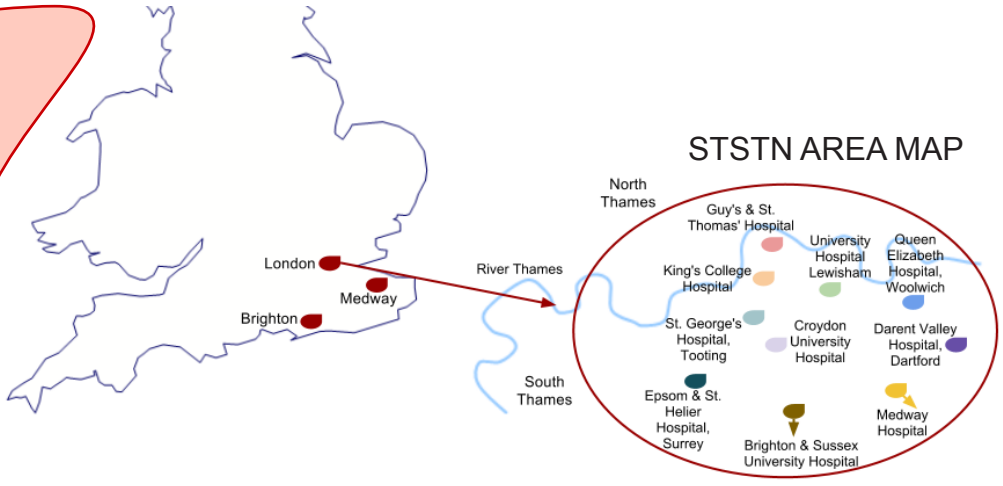
STSTN is currently funded by King's College Hospital Charity, the UK Forum on Haemoglobin Disorders and Roald Dahl's Marvellous Children's Charity.

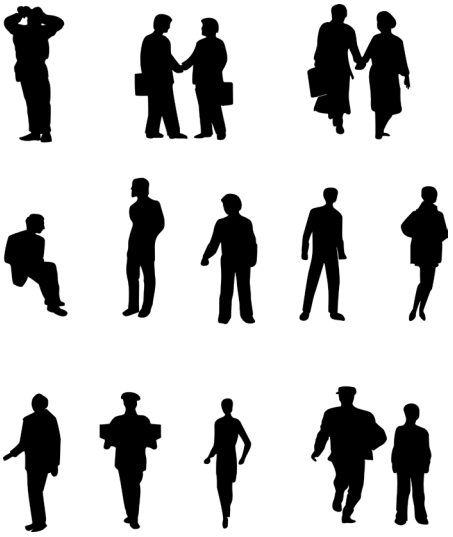
Read about STSTN's Key Objectives on page 3....

STSTN'S MISSION

"To improve the patient experience by offering better treatment outcomes for people with sickle cell disease and thalassaemia"

STSTN AREA MAP





PATIENT PLATFORM

YOUR STORIES. YOUR NEWS. YOUR VOICE.

BY AIMY A.

Somebody once told me that your school days are the best days of your life. For children with sickle cell disease "best" may not be the correct descriptive word. While all my friends were going to end of year parties, camping trips or foreign exchanges I was confined to the exciting world of the hospital. My idea of travel was being air lifted from hospital to hospital. I had to make the best of not only a limiting and chronic disease, but the physical change and emotional turmoil we face as teenagers.

There was an all-consuming fire burning inside me. I was angry at my blood, God, my parents for creating a defective child, the world for making it so difficult for me to succeed, at the nurses' open insinuations that I was a junkie because I was black and needed to take opiates. I grew up not knowing anyone else with sickle cell and had no one to talk to. I felt I had to stick on a strong face and not admit that just under the surface I wasn't just angry, I was scared. My world was dominated with people that had a lot of sympathy and text-book knowledge. Empathy, however, was in short supply. This made me feel sharing my feelings would be pointless.

I survived my angry teenage years and entered my twenties. I grew up in a lively family and developed a strong sense of my self and my independence. I was constantly out for adventure but years of hospital trips left me feeling confined. I wanted to get out and explore, not just the world but myself. I needed to test my limits and find an identity beyond sickle cell. I craved validation and to find out what makes life worth fighting for. So far I had only experienced fighting to be well and waiting to be sick. My parents had never put pressure on me to attend university; just staying alive was enough for them.

I decided to start working. I joined a temp agency and secured employment working as an administrator for Barclays plc. and the DWP. I left my parents' house and into rented accommodation with my boyfriend. My new found freedom was fabulous! I was young and in love. It was good times until my crises started getting closer and closer together.

Losing chunks of my life to sickle cell made it almost impossible to keep my job. No job meant the end of independence. Goodbye happiness, hello hospital. I felt completely alone, I was signed off work and had moved back home. Being sick officially sucked. I drifted through life, moving from place to place, doing nothing and feeling unfulfilled and unsettled. I needed a change. I needed to stop dwelling on the negative and to not allow myself to fall into the trap of being defined my illness. For my own sanity I had to be brave and make changes.

I split up with my boyfriend and started taking Hydroxyurea, a drug that raises foetal haemoglobin within the blood. For reasons unknown, I was initially strongly against taking Hydroxyurea, but it turns out it was the best thing I ever did. Within months I felt better, I had more energy and the aches and pains I had become used to ignoring slowly disappeared. I took full advantage of my sudden good health. I left everyone I knew and loved in Hastings and moved to London alone. I started the adventure with the mantra "I'm going to live a little before I die!" and never looked back.

Now, in my mid-twenties I have learnt not to get anything get me down. Of course I have to work within my limits, but it's been quite fun pushing those limits. Do not be afraid to challenge yourself. If you want to party – go for it! If sport or education is more your thing, pursue it! Being the next Donald Trump is not out of your reach. It is attainable if you want it.

The little girl that was constantly told that she couldn't or wouldn't have a "normal" life, point blankly refused to let sickle cell or anything else hold her back. Sometimes when it all feels like an uphill struggle, I remember all the battles I've won. I remind myself of that stubborn little girl and I realise that I can do it, I can rebuild physically and mentally one step at a time. I've worked, studied, partied and travelled just like any other twenty-something. It's not easy but I would not trade my life for the world. The lessons I learnt as a child prepared me for the knocks of life and I realised that you cannot win if you do not play the game. Get off the side-lines and TRY. Don't ever give up! If I can do it, so can you!!

Aimy A. is currently a patient at King's College Hospital

PATIENT PLATFORM

YOUR STORIES. YOUR NEWS. YOUR VOICE.

BY SUSAN FLEMMING

EXPERT PATIENTS PROGRAMME

Susan Flemming, a sickle cell patient at Guy's and St. Thomas' Hospital NHS Foundation Trust, talks about her role in the Expert Patient Programme.

The Expert Patients Programme (E.P.P) is a free self-management course that gives you the tools to enable you to take control of your long-term health condition. It's a 6-week group course, attending 1 day a week for two and half hours per session, with two tutors facilitating the course who also live with long-term health condition/s.

Whilst on the course I learnt ways of dealing with the symptoms that come with having a long-term health condition, for example, tense muscles, stress and anxiety, pain, anger, fear and frustration. An example of using the self-management tools is that when I felt stressed and was having a bad day I would use the relaxation and better breathing techniques to help release the tension I was feeling that day. I learnt how to deal with my anger, fear and frustration by communicating with friends, family and with the health care professionals, also using positive thinking to help motivate myself when thinking negative thoughts. Another great tool was learning to pace myself by managing my day effectively by planning and making an action plan.

Action planning has become a major part of my life today as it allows me to note if I am doing too much on a day-to-day or weekly basis. If my day is not managed correctly I will more often than not trigger off symptoms of the illness or endure a flare up. It is also a great way of keeping track of any long or short-term goals I may have set for myself.

I have been an Expert Patients Programme Tutor for 8 years now, facilitating courses in many areas of London. Becoming a tutor has helped me not only to be in more control of the illness, but to also acknowledge when I need to take things easy. It has also allowed me to share and help many others achieve the same goal in not allowing their long-term condition to control them but for them to take control of it. Many of us who have a long-term condition/s tend to do all that we can in one day and therefore over doing things which then can makes us unable to do anything the next day. I truly believe in this course and would like to encourage sickle cell patients as well as anyone with a long-term health condition/s to join a course today. Do not hesitate to call your local Primary Care Trust today and take control.

For further details:

Expert Patient Programme can be found on the NHS wesbite: www.nhs.uk/conditions/Expert-patients-programme-/Pages/Introduction.aspx

STSTN's Key Objectives

SERVICE

- Establish and maintain joint methods of working throughout the network
- Collaborate in specialist clinics including renal, obstetrics, orthopaedics, cardiology, neurology, urology and liver.
- To publish guidelines on the care and treatment of sickle cell disease and thalassaemia
- To produce patient leaflets and separate patient newsletter

TEACHING

- To increase the understanding of a patient's experience of SCD and thalassaemia via Patient Forum/Awareness events
- To provide a peer-support network and educational programme for consultants, GPs, nurses, trainees and other healthcare professional

RESEARCH

- To disseminate information and raise awareness of ongoing research in the UK and internationally
- To function as a group and provide critical mass for collaborative research and evidence based audits

Dr Baba Inusa
Consultant Paediatrician at Evelina Children's Hospital

AN UPDATE ON THE SILENT CEREBRAL INFARCT MULTI-CENTER CLINICAL TRIAL

Silent infarct transfusion trial, otherwise known as SITT for short, is a phase 3 trial to investigate the role of blood transfusion in brain injury or silent cerebral infarct (silent stroke) in children with sickle cell anaemia.

In sickle cell anaemia, stroke may occur at any anytime, it may be overt (associated with neurological deficit) or silent (no obvious neurological deficit) but a well defined lesion on brain MRI scan. Silent stroke therefore may only be detected using brain MRI scan. Whilst the prevalence of overt stroke occurs in about 11 out 100 children by age 18, silent stroke occurs in about 25 out of 100 children.

Goal of study

To determine the effectiveness of blood transfusion therapy for the prevention of silent cerebral infarct (stroke) in children with sickle cell anaemia. It will also determine if blood transfusion will prevent further brain injury; whether the benefits of blood transfusion outweigh the risk.

This is phase 3 trial intervention trial

Primary outcome: The development of progressive cerebral infarct assessed by MRI of the brain or overt stroke
Start date: December 2004, project end date: May 2013 and data collation is December 2013.

Age group

5-14 years with sickle cell anaemia (HbSS or Sickle Cell beta zero- Sβ), both boys and girls are included.
Intervention arm / transfusion group receive regular blood transfusion 4-6 weekly to maintain the level of haemoglobin ratio at 30% or less, over a 36-month period.

The study was approved by ethical institutions and the research and development departments of all participating institutions. It is funded by the US national institute of health to a tune of $20million dollars. The chief investigator is Professor Michael DeBaun while at Washington University Hospital, St Louis and now at Vanderbilt University, Nashville. Together with the deputy co-chair Professor James Casella of Johns Hopkins, Baltimore and over 26 experts investigators across the world - USA (21 centres), France (1), UK (3) and Canada (1).

Silent Infarct Transfusion Trial

Study sites in the United Kingdom

Three UK centres recruited participants into the study:Evelina Children's Hospital (GSTT), The Royal London and Bart's Hospital and Central Middlesex Hospital, all London institutions. The investigators in the UK are Professor Fenella Kirkham, Dr Paul Telfer , Dr Baba Inusa, Dr Michele Afif and Dr Kofi Anie

What does being on the study entail?

Only those who were fully informed of the study aims and understood their rights to choose and decided to give their consent (adults with parental responsibility) were included.

The participants are initially screened using brain MRI scan to determine evidence of brain injury (silent stroke), which is not associated with an overt stroke. If the initial screening scan is positive for silent infarct, a second level of consent was required to go to the next stage MRI scan.

The randomisation was done at the central control site in the USA. In effect neither the doctor nor patient could determine which arm of the study the child is allocated to. Each arm of the study is expected to last 26 months except if the child reaches an end point i.e. attained one of the outcome of the study.

When do we expect the final outcome?

The last study completes the trial on the 31st May 2013 and the final data collection is December 2013. Already a number of publications have taken place including the contribution of hypertension on the risk of brain injury in sickle cell anaemia. These and future publications are providing useful understanding for the management of sickle cell anaemia. These may in fact change the long term management of children with the disorder.

HEALTH PSYCHOLOGY SERVICES

Dr Nicky Thomas,
Clinical Health Psychologist, Guy's & St. Thomas' NHS Foundation Trust

VOLUNTEER FOR A STIGMA RESEARCH STUDY

STUDY AIMS

To determine the extent to which 'felt' and actual stigma exisits today in sickle cell disease (SCD) patients in the UK and to ascertain how this impacts on their health and accessing health care.

BACKGROUND TO STUDY

Stigma is the discrediting, degradation and discrimination of individuals who are perceived to possess attributes that are disparaged in society (Marks et al, 2005). It is a form of social oppression that prevents a group's full participation in society. There are two main types of stigma; 'felt', the feelings associated with having a stigmatising illness and 'actual', the discrimination that the individual encounters (i.e. the denial of jobs and access to health care). SCD largely affects people of African descent and historically, people from this group have experienced stigmatisation based on assumptions about their race, character and identity. In SCD there are many elements that contribute to making it a 'felt' as well as an 'actual' stigmatising condition. These include genetic heritability, notions of contagion, blood borne illness, impact of illness causing disability and the stigma emanating from this feeds learnt helplessness. For some people with SCD, they are at risk of further stigmatisation due to factors such as their need for opiates, being unemployed and the experience of psychological consequences. The 1960's and 70's saw SCD being largely transformed from it being a stigmatising illness to a worldview of it being a painful disease. Since then, the transformation of sickle cell disease has continued and, in the 21st century, there is increased understanding gained through biopsychosocial research.

Yet in spite of this there is still a strong element of interplay between racial and disease stigma but other than anecdotal information there is very little evidence identifying the extent of felt or actual stigma in sickle cell patients in the United Kingdom. It is suggested that stigma could interfere with necessary treatment in a number of ways:

- It could delay people seeking treatment
- Efforts to hide the diagnosis from others could interfere with adhering treatment.

Therefore the objective of the present research proposal is to determine whether 'felt' and 'actual' stigma exists today in SCD patients. We also seek to ascertain from patients their views about how health psychology input could help to address stigma if it is identified.

OBJECTIVES OF RESEARCH HYPOTHESIS

1.To ascertain the extent of stigma in patients with sickle cell disease.
2. To describe the factors that patients consider to be stigmatising
3. To ascetain what kind of intervention would be helpful to reduce the level of stigma

For more information on the study, volunteering and collaboration, please contact Nicky via email: nicky.thomas@gstt.nhs.uk or phone: 020 7188 2725

Did you know...?

It's Guy's and St. Thomas' Sickle Cell Awareness Day on 12 July 2012

King's College Hospital's Patient Forum day is on 13 July 2012

You can email your comments and content suggestions for red cell *news* to: info@ststn.co.uk

SICKLE CELL AND THE BRAIN

Dr Heather Rawle
*Clinical Health Psychologist, Guy's & St. Thomas' NHS Foundation Trust*st

NOW WHERE DID I PUT THAT APPOINTMENT LETTER?

Most people with sickle cell disease go through their lives without any problems with their brain but we know for some people, sickle cell can affect the brain, just like it can affect any organ of the body. As a result, some people may notice that they are more forgetful, lose concentration more easily, or feel less able to keep organised. Such problems are called 'cognitive impairments' and we know they can also happen when someone is under a lot of stress or feeling particularly low in mood. For adult patients who receive their sickle cell care at Guy's and St. Thomas' NHS Foundation Trust (GSTT), the Health Psychology Service can do a cognitive assessment to look at how someone's memory, concentration and problem solving skills are. It shows what your strengths and limitations are, so we can work with you to find ways of coping. This can help you in your everyday activities, working life or studies. The assessment is made up of several spoken, written and puzzle-type tasks and takes around four hours to complete.

At GSTT we have looked at the results of our adult patients who had cognitive assessments and MRI brain scans. Some of these patients had a stroke (an interruption to the blood flow in the brain), some had silent strokes (where the interruption is so small the person is not aware it has happened but it shows up on the brain scan) and others had no past strokes or silent strokes. We found that patients tended to have problems in the following areas: the speed with which the brain can take in information, concentration, and skills that are important for problem-solving. These problems were worse for people who had strokes but were also present to a much lesser degree in patients who had silent strokes. These types of problems can explain why some people are forgetful. This may include forgetting appointments, forgetting to take medication, and finding it hard to keep on top of studies or work. We found that doing cognitive assessments was helpful in finding out about these problems which meant we could give patients guidance about how they can build on their coping strategies. It also means that we can help hospital staff to understand more about how they can help patients.

As mentioned earlier, most people get through their lives without any effect on their brain but if you would like to know more about how sickle cell can affect the brain, then speak to your sickle cell Doctor.

Five Top Tips for Improving Memory

- Use a calendar/phone for appointments
- Keep a to-do list
- Write things down that you need to remember later
- Keep things in the same obvious place so that you can locate them easily e.g. keys by your door
- When you need to concentrate, get away from distractions

DON'T FORGET...GET INVOLVED

If you *have a story to share or would like to offer your design, illustrating or photographic skills to red cell news please get in touch.*

Likewise if you have any events you would like publicised that connects sickle cell and thalassaemia patients and their families, do let us know.

Email info@ststn.co.uk or call 020 7848 5441.

With thanks to our funders

King's College Hospital Charity · UK Forum on Haemoglobin Disorders · Roald Dahl's Marvellous Children's Charity – Making life better for seriously ill children · NOVARTIS